Journal of Pediatric Surgery Case Reports 69 (2021) 101856

Contents lists available at ScienceDirect

Journal of Pediatric Surgery Case Reports

journal homepage: http://www.elsevier.com/locate/epsc

Inflammatory myofibroblastic tumor of head of pancreas in a 5 Year-Old child

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ARTICLE INFO

Keywords:
Inflammatory myofibroblastic tumor
Pseudotumors
Pancreas tumors

ABSTRACT

Inflammatory myofibroblastic tumor (IMT) is assumed as a rare benign tumor that can appear in various organs. Since Umiker et al. introduced inflammatory pseudotumors for the first time in 1954, it was found that occurrence in the head of pancreas is very rare (Baião et al., 2019) [1]. Differentiating non-neoplastic lesions of pancreas from adenocarcinoma remains a challenge despite advances in diagnostic modalities whereas cystic and solid lesions and some normal anatomic variants can mimic malignancy (Okun and Lewin, 2016 Jan 1) [2]. Histologic patterns of IMT can be identified as fibroblastic and myofibroblastic proliferation with inflammatory infiltrate. They are more common in Women and peak of occurrence is between 2 and 16 years of age (Mirshemirani et al., 2011) [3]. In head of pancreas the disease can present with obstructive jaundice and the key for diagnosis is having a tissue specimen by means of needle biopsy. Though in all reported cases surgery is suggested as the lone treatment option, its natural history is obscure still and there can be a challenge in its treatment and also the extent of surgery.

Here a new case of IMT of head of pancreas is reported.

1. Introduction

Inflammatory myofibroblastic tumor is a rare disease, at the same time an interesting phenomenon in regard to its etiology, presentation, diagnosis and treatment like many other rare conditions.

Lung, mesentery and omentum are the most common sites of IMT occurrence, and besides them, in other organs like: Gastrointestinal tract, liver, abdominal soft tissue, spleen, lymph node, and extremely rare in the pancreas [4,5].

Here, we report a 5 Year-Old patient with chief complaint of jaundice that after further investigations turned out to be Inflammatory myofibroblastic tumor of head of pancreas.

2. Case presentation

The patient is a 5 year old female with the chief complaint of yellowish discoloration of sclera in December 2019. She was doing well until she developed progressive jaundice for duration of one month. Except for a minimal loss of appetite, she was not suffering from any other symptoms such as weight loss, nausea, vomiting or abdominal pain. She had neither previous gastrointestinal problems nor significant past medical history.

On physical examination only icteric sclera and icteric skin were detected. Abdomen was completely normal on examination. Laboratory investigation showed a rise in ESR (59mm/hr), high bilirubin (Total: 4.36, Direct: 2.85mg/dl) and elevated liver enzymes (AST: 232 U/I, ALT: 312 U/I, ALK P: 2596 IU/L).

Sonographic findings revealed dilated CBD (9mm) and interahepatic bile ducts. A hypoechoic mass in pancreatic head measuring about 22*22*19mm without vascular invasion was noted. Besides, Pancreatic duct was mildly dilated. Contrast-enhanced MDCT images demonstrated a 22*21 mm mass in the head and uncinate process of the pancreas. After contrast administration, its appearance showed a hypodense lesion. There was not any metastatic foci in the liver. The superior mesenteric vein and artery were intact with normal calibre. Axial pancreatic-phase MDCT (E) displayed the “double duct” sign, consisting-of common bile duct and pancreatic duct dilation. Also, first generation of interahepatic bile ducts dilation and abrupt distal part of common bile duct and pancreatic duct obstruction were noted (Fig. 1).

Following the above investigations a CT guided needle biopsy was performed. The first review of the tissue was inconclusive; consequently we tried prednisolone with impression of IgG 4 related disease and autoimmune pancreatitis. After about one month treatment, there was no any response either clinically or paraclinically, though the pa-

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https://doi.org/10.1016/j.epsc.2021.101856
Received 4 March 2021; Accepted 28 March 2021
Available online 8 April 2021

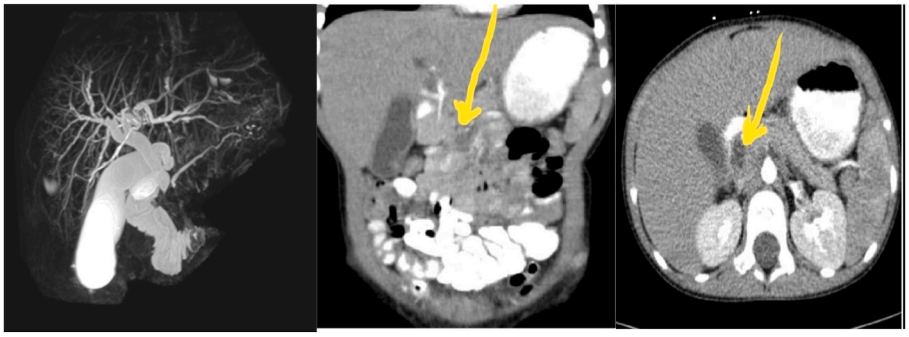

Fig. 1. Radiologic findings
Contrast enhanced CT scan of the patient reveals an ill-defined 28*25mm solid mass in the head of the pancreas with a dilated common bile duct — the " double duct " sign. Also interahepatic ducts are dilated.

tient's condition did not deteriorate. Further review of the same tissue specimen led to another diagnosis: Histologic findings demonsterated an inflammatory myofibroblastic tumor of pancreas. Also storiform and fascicular growth of spindle-shaped and ovoid cells with prominent inflammatory infiltrate was evident (Fig. 2).

Following this new finding we came to the conclusion that the patient should undergo whipple operation according to a multidisciplinary meeting decision. The wipple operation constituted of the classic pancreaticoduodenectomy with lymph node dissection. Operative findings were a hard 3*2 cm mass inside the head of pancreas with mildly dilated common bile duct; otherwise other organs were normal. The Post-operative course was uneventful; there was only chyle drainage from the abdominal drain catheter that was self-limited and ceased after a few days. The patient tolerated the operation well and was discharged on the 7th postoperative day with good general condition. Surgical pathology study confirmed preoperative diagnosis of IMT of head of pancreas with the greatest dimension of 2 cm (2*2*1.5cm). (Fig. 3).

The removed lymph nodes were not involved by the tumor. On the 6th postoperative month follow up she had developed diarrhea after consumption of fatty meals, laboratory data including AST, ALT and bilirubin were normal, her stool elastase was low (=43micg/ml) and she is on pancrelipase (CREON); otherwise she feels well. Other laboratory data including: Amylase, lipase, Gamma GT, Serum Albumin and cholesterol were within normal limits, but Triglycerides was borderline and 25-OH vit D level was insufficient. .After one year follow up, now she is alive and has good general condition with normal liver function test.

3. Discussion

Inflammatory myofibroblastic tumor is considered as a benign inflammatory lesion which develops in the soft tissues. There are a variety of names used to describe IMT such as: plasma cell pseudotumor, plasma cell granuloma, histiocytoma, inflammatory pseudotumor and inflammatory fibroxanthoma [6].It usually is detected in the first two decades of life, and females are more affected [3]. Although usually asymptomatic, they are incidentally detected in a routine evaluation in imaging or laboratory tests, but depending on the site of involvement signs and symptoms can be different [7].According to a literature review which encompasses all similar cases (27 cases):Abdominal pain, jaundice, anorexia or weight loss, and nausea or vomiting are more frequent symptoms respectively [8].

In another study of 32 patients under 18 years of age, conducted by Dalton et al. it was shown that the abdomen was the most common site of occurrence. Involvement of the mesentery when the tumor location is in the abdominal cavity is usual. Colon, appendix, esophagus and stomach are other reported sites of involvement [3,9]. Mir, Mohamad Hussain et al. reported in a study of pseudotumors where five out of 288 pediatric tumors diagnosed as IMT [7]. According to our best knowledge six cases of this kind of mass lesion in the -head of pancreas are reported in children under 18 and the youngest being a six month old child [10]. AlirezaMirshemirani et al. demonsterated four Iranian pediatric cases between (3.5–13years), diagnosed Abdominal IMT in different organs (stomach, periduodenal, mesenteric and colon [12].

Taken together, our patient is first case repor of pediatric pancreatic IMT in Iran. As an uncommon neoplasm of children, IMT rarely has the Potential of malignancy [9].

Due to common mistakes for diagnosis of IMT versus pancreatic cancer, tissue specimen is needed for definite diagnosis because almost always it is made by using histopathological methods [1,2,11]. Fine needle aspiration can rarely be helpful for diagnosis and inflammatory markers are not appropriate and reliable enough to rule out IMT.(10 (

Myofibroblastic and fibroblastic spindle cells with inflammatory infiltrate of lymphocytes, histiocytes, plasma cells and eosinophils may be

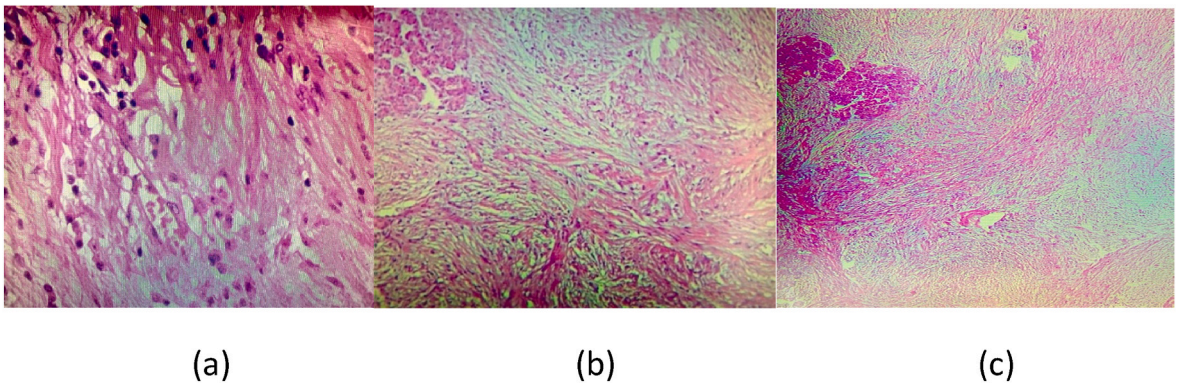

Fig. 2. a&b. Low power showing storiform and fascicular growth of spindle-shaped and ovoid cells with prominent inflammatory infiltrate.
c. High power; note the conspicuous admixture of lymphocytes and plasma cells.

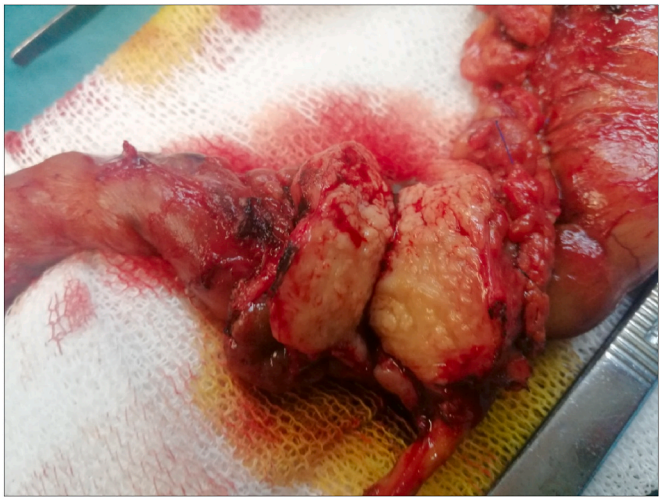

Fig. 3. Macroscopic view of the resected parts.

seen in histologic investigations. In a gross view, they are circumscribed but not encapsulated and may have focal parts of necrosis, calcification or hemorrhage [7].

Although there are some reports of lesion regression but due to its similarity in presentation to other tumors and lack of clear guidelines in the treatment of IMT, surgery was utilized as the first line treatment. Despite the chance of recurrence after surgical resection and reported cases with mortality and distant metastasis which named inflammatory fibrosarcoma, IMT considered as a benign lesion [3].Nevertheless the natural history of the disease as a whole and those involving a special organ like pancreas is not precisely known. Therefore the extent of surgery including lymph node dissection and adjuvant therapy are the issues that need further clarification in future.

The diagnosis of more cases and long term follow up in a multidisciplinary setting remains the sole way to determine the unknown clinical aspects of this disease.

Consent

Consent to publish the case report was not obtained. This report does not contain any personal information that could lead to the identification of the patient.

Funding

There is no funding or grant support for writing this paper.

Authorship

All authors attest that they meet the current ICMJE criteria for Authorship.

Declaration of competing interest

The authors declare that they have no known competing financial interests or personal relationships that could have appeared to influence the work reported in this paper.

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